JUVENILE XANTHOGRANULOMA*
REPORT OF A CASE
BY
J. GIBSON MOORE AND JOHN HARRY
Chase Farm Hospital, Enfield, Middlesex
and the Department of Pathology, Institute of Ophthalmology, University of London

JUVENILE xanthogranuloma, also commonly known as naevoxantho-endothelioma, was first described as a skin disease in 1909 (McDonough, 1909), but it was not until forty years later that the first intra-ocular case was reported in America (Blank, Eglick, and Beerman, 1949). Some twenty to thirty cases have since been described (Sanders, 1962), but our case is the first to be reported in the United Kingdom.

Case Report
A 12-month-old boy was first seen on January 12, 1962. The parents stated that one week previously they had noticed a yellow mass in the pupil of the right eye.

Examination.—There was a large mass on the iris extending from 3 to 7 o'clock involving the pupillary margin together with an area of vascularization at the edge of the pupil, but no sign of haemorrhage into the anterior chamber (Fig. 1).

![Fig. 1.—Right eye of patient showing iris tumour in lower nasal quadrant.](http://bjo.bmj.com/)

Examination under General Anaesthesia.—The cornea was seen to be clear and the fundus normal, but gonioscopy revealed that the mass on the iris was spreading into the filtration angle. At that time the intra-ocular pressure was 16 mm. Hg (Schiötz). Apart from the presence of this iris tumour the child appeared healthy and no skin lesions were detected.

Operation.—A tentative diagnosis of malignant iris tumour having been made, the lesion was removed by performing a large iridectomy on February 12, 1962. During the period of one month which had elapsed since the child was first seen the tumour had enlarged, and at the time of operation it occupied approximately one-third of the iris. From a technical viewpoint the operation was

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difficult; not only was the tumour in the lower nasal quadrant of the iris, but it was friable and could only be removed by a radial incision into healthy iris tissue on either side and cutting across the root. There was considerable haemorrhage at the operation, at the conclusion of which there was a total hyphaema. The wound was closed with three corneoscleral sutures. It was considered that removal had not been complete and that tumour remnants remained in the angle of the anterior chamber.

Post-operative Course.—During the first few days after the operation the hyphaema appreciably lessened in amount, but the cornea became oedematous and the intra-ocular pressure rose to 48 mm. Hg (Schiotz).

Pathological Report.—Sections of the tumour showed the iris stroma to be diffusely infiltrated with monocytic cells (Fig. 2), some of which had a clear eosinophilic cytoplasm, while others had a foamy lipid cytoplasm giving rise to a vacuolated appearance (Fig. 3). Several typical Touton giant cells (Fig. 4) consisting of a lipid cytoplasm with a ring of nuclei were seen, together with an occasional foreign body giant cell (Fig. 5). A number of small blood vessels
was present (Fig. 6) and there was a little haemorrhage in addition to some necrosis (Fig. 7). The histological appearance was that of xanthogranuloma.

**Follow-up.**—On February 24, 1962, the patient was referred to Dr. V. B. Levison at the North Middlesex Hospital for radiotherapy, and two doses of 200 r were given at an interval of 7 days. The result of this treatment was quite dramatic. After 10 days the corneal oedema disappeared, the eye became white, and the iris pillars free. There were no lens opacities, but a circular ring of pigment, the size of the normal pupil, was observed on the anterior surface of the lens.

In view of the diagnosis a further search for skin lesions was made but none was found. The following investigations were performed: X-ray of skull and long bones—no abnormality detected; blood count—normal; blood cholesterol—240 mg./100 ml.; urine examination—negative.

It is now over two years since the lesion was removed. The eye has remained quiet and there has been no recurrence of the tumour (Fig. 8).
is, however, unknown. The anterior surface of the eye is a complex structure consisting of multiple layers and components, each playing a crucial role in the function of the eye. Localized synechiae, for example, can occur when the iris adheres to the anterior lens capsule, often following injury or inflammation.

Histologically, there is often a similarity between the involvement of the skin and the eye. The skin lesions may appear as xanthogranuloma, a histiocytic reaction that is characterized by the presence of lipid-containing histiocytes. These cells may accumulate lipid in the cytoplasm, forming lipid droplets that are visible under the microscope. The histological picture of the skin lesions is often striking, with a marked infiltration of monocytes which often have polygonal fat-storage disorder. It is believed that this infiltration can extend into the trabecular meshwork and ciliary body, causing a rise to intra-ocular involvement in cases of juvenile xanthogranuloma.

Secondary glaucoma may also be seen, with resultant rise to intra-ocular pressure and resultant loss of vision. It is a condition that is seen most frequently in children, occurring either congenitally or from an injury. Secondary fibrosis has been seen in cases of intra-ocular involvement, leading to fibrosis of the iris and ciliary body. This condition is relatively rare, but when it does occur, it is often associated with a history of radiation therapy to the head or scalp.

Local radiation to the head and scalp is known to cause fibrosis of the iris and ciliary body. However, there does appear to be an increased incidence of xanthogranuloma in the anterior chamber in children, and this condition is often associated with other systemic disorders, such as sickle cell disease. In general, the presence of xanthogranuloma in the anterior chamber is an indication of a systemic disorder, and appropriate investigations should be undertaken to determine the underlying cause.

The skin is a highly characteristic organ of the body, and it is known to exhibit a number of unique features that distinguish it from other body tissues. One of these features is the presence of the pigment melanosomes, which are produced by the melanocytes of the skin. These cells are responsible for producing the pigment melanin, which is responsible for the coloration of the skin. In addition, the skin is also known to be highly sensitive to various external stimuli, such as temperature, light, and mechanical trauma. This sensitivity is reflected in the ability of the skin to respond to both acute and chronic stimuli through a number of different mechanisms, including vasodilation, sweating, and abrasion.
lesion with hyphaema and/or secondary glaucoma. In order to prevent unnecessary enucleation it is of considerable importance to make the correct diagnosis, and this can be established either from the typical clinical picture or from the histological appearance of skin or iris biopsy.

Summary

A case of juvenile xanthogranuloma in a male infant aged 12 months is reported. The pathology and possible pathogenesis of the lesion are discussed.

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REFERENCES

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